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A CASE OF CORTICAL SCLEROSIS, HEMIPLEGIA AND EPILEPSY, WITH AUTOPSY.*

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Summary: Child, age four and one-half, forceps delivery, right hemiplegia from birth, general epilepsy from sixth month, operation, death, meningeal thickening, general cortical sclerosis in area of distribution of Sylvian artery, atrophy, degeneration of pyramidal tract, no degeneration of lemniscus or atrophy of cerebellum, considerable deficiency of tangential fibers.

The case which I have here to report furnishes a contribution to the pathological anatomy of infantile hemiplegia with epilepsy and helps to throw light upon the origin of at least some cases of this disease.

Leo L., age 4½ years, Hebrew, born in the United States, was referred to me by Dr. George Woolsey and I am indebted to him for the opportunity of seeing the case and of using the material. The family history is quite negative. The child was the first born and the labor was practically difficult, forceps being used. The infant was alive and had no convulsions at this time. There was no defect noticed at first, but within a few months afterward it was found that he did not move the right arm and leg quite so much as the left. He was a bottle baby and was poorly nourished, almost marasmic. The fontanelles were large and slow in closing and the mother says that the face was always bluish. During the second year of life he became quite healthy and looked well, but never used his right hand very much. The child began to walk at the age of two years, and talked a little at the end of the first year. Teeth first appeared at nine months, at which time he suffered from a bad diarrhea. When two years old he began to have convulsions without known cause, and these continued during the rest of his life, but changed in their character. At first the attacks consisted of choking sounds, the eyes turned to one side and there was a blueness of the face and stiffening of the limbs. This was followed by complete loss of consciousness for about a quarter of an hour with clonic convulsions, each side being affected. There was some vomiting. After this he would lie in a stupor for one or two hours. Later on the convulsions became less severe and they were characterized by smacking of the lips, irregular gestures and stiffening of the joints and of the body. No Jacksonian type was ever noted. He suffered a good deal from headache, and as he grew older he showed considerable backwardness in intelligence.

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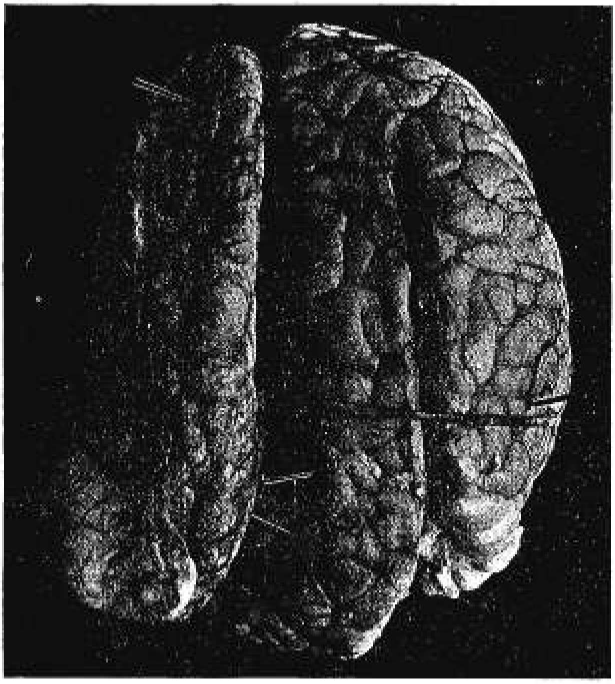

Fig. I. View of convexity of the hemispheres.

When seen by me at the age of 4½ years he was a well-nourished boy about normal height and weight. In walking there was a slight hemiplegic gait, and examination showed a distinct right hemiplegia involving the arm mostly. The right foot was somewhat stiff and had a tendency to turn in. The right forearm, the wrist and the fingers were flexed and were decidedly rigid. There was exaggeration of the reflexes upon the right side. The head measured 48 ctm. in its great circumference; there was some facial asymmetry. He could not use the right arm very well, and could not pick up a penny with his right hand, on account of the weakness and rigidity of the parts. Electrical reactions were normal in all the muscles except the forearm, where they were diminished. Facial innervation was less upon the right side. The right leg showed no shortening nor atrophy, but there was some shortening and atrophy of the right upper extremity. He had no athetoid or clonic movements. All the spinal reflexes were present, except the abdominal. The plantar reflex was of the flexor type. He could talk a little, but not very much. There were no disturbances of sensation or of the special senses. For the details of this examination I am indebted to Dr. Saml. M. Evans.

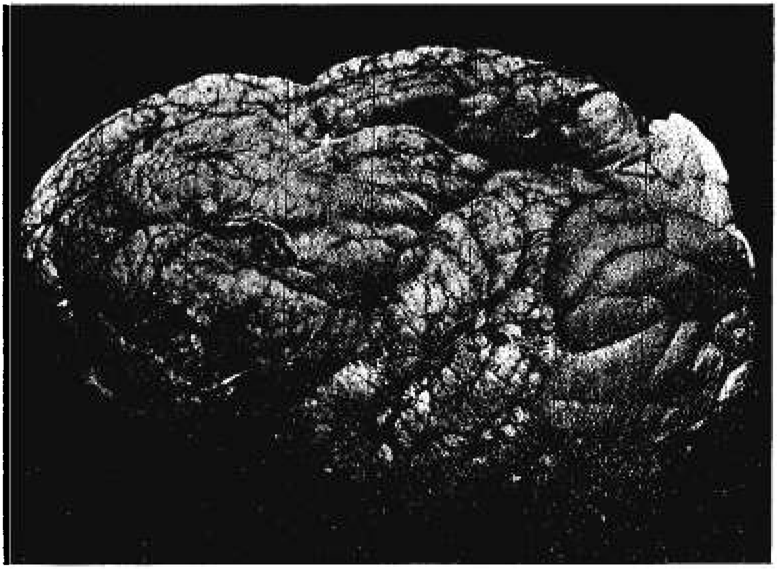

Fig. II. Lateral view of left cerebrum showing microgyry, thickened pia and normal occipital lobe.

He was having the epileptic attacks with increased frequency, and owing to this fact and the hopelessness of medication, as tested by previous experience, I advised Dr. Woolsey to make an exploratory operation, hoping that, as sometimes occurs, some relief from the epileptic symptoms might be obtained through the emptying of the cyst and the removal of pressure, or through some mechanical change in the circulation. Dr. Woolsey therefore trephined and found the atrophied brain lying under the opening. He opened the lateral ventricle, which was distended, and withdrew some fluid. The child did not survive the shock of the operation. A post-mortem was made and the specimens were preserved and sections made by Dr. Schlapp, to whom I am indebted for the preparations. The photographs of the brain show better than any description the conditions found.

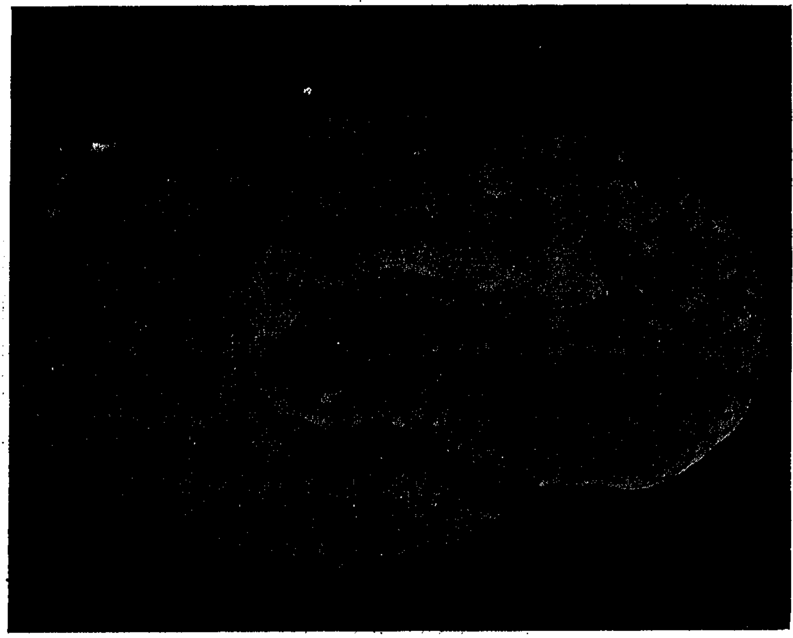

Fig. III. Median surface of cerebrum.

Report of Autopsy.—The left hemisphere of the brain is greatly atrophied, being about one-third smaller than the right. The atrophy involves all the cortex except the occipital lobe, and to a less extent the anterior part of the frontal, which is normal. The atrophy corresponds very nearly to the distribution of the Sylvian artery. It involves the convolutions, which have shrunk up so that the typical fissures cannot be made out, except the Sylvian, and those of the lower temporal, occipital and anterior frontal lobes. The small gyri are bound together by thickened connective tissue developed from the pia-arachnoid; and the vessels running into them are often obliterated. There is, however, no diffuse or lobar sclerosis. The increase of connective tissue being due simply to proliferation from the vessel walls and the pia-arachnoid. The condition is one of microgyry due to vascular defect. There is no evidence of venous thrombosis. The lateral ventrical is considerably dilated (twice its normal size).

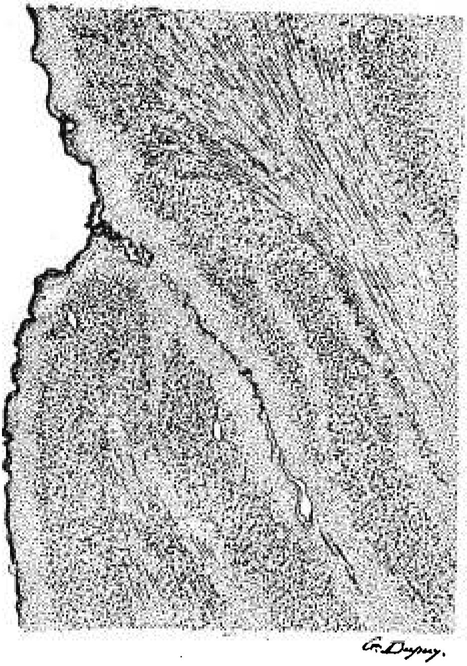

Fig. IV. Section of prefrontal convolution, showing partly obliterated fissure and areas of diffuse sclerosis. ×1½.

Microscopical examination (Nissl, Delafield, Weigert stains) shows some thickening of the pia, but no true meningitis. The vessels are not diseased, but many of the small pial arteries running into obliterated fissures are obliterated.

Throughout the cortex one sees areas of sclerosis which lie around the pial vessels, and which are evidently due to obliteration, partial or complete, of these. There is no independent sclerosis, and the trouble is plainly secondary to the vascular defect. The sclerotic areas are very numerous, but do not extend into the white matter far, and the white matter is normal, so far as this process is concerned.

The cortical nerve cells are degenerated in many places, being small, irregular in shape and deficient in number. Many areas are seen in which the cells are fairly normal. It is impossible to make out any tangential fibers in most areas.

The fibers of the subcortical tissue show degenerative changes, but the study of these as yet is not complete. The basal ganglia were not affected. The cerebellum shows no atrophy. Sections through the pons and medulla show the usual degeneration of the pyramidal tract, those bundles near the median line in the pons being most affected. The atrophy involved about three-quarters of the tract on the right side. The posterior longitudinal bundles were normal. The lemniscus was not atrophied on either side. These findings are in harmony with other observations to the effect that when the

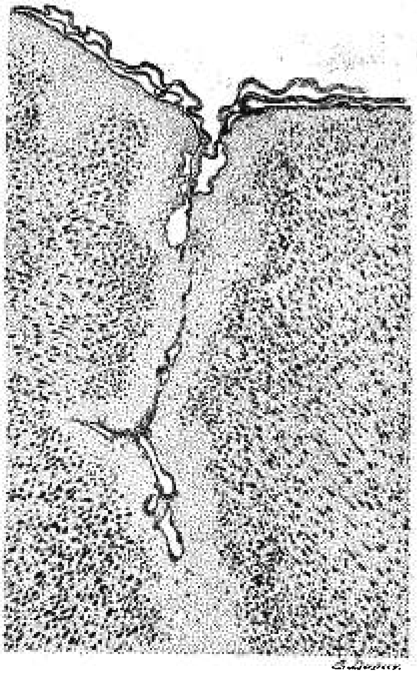

Fig. V. Section of prefrontal convolution, showing fissure with sclerotic areas and degeneration of cells. ×⅔.

lesion is cortical there is no degeneration of the lemniscus or atrophy of the cerebellum.

Remarks: The lesions which are found causing conditions more or less similar to these clinically, that is to say, hemiplegia or diplegia and epilepsy are the following: 1. Diffuse sclerosis. 2. Lobar sclerosis. 3. Atrophic sclerosis. 4. Hypertrophic sclerosis. 5. Porencephaly. 6. Agenesis. 7. Cysts. 8. Simple atrophy. These various anatomical defects have as a cause (a) simple developmental defects, (b) chronic hydrocephalus, (c) meningo-encephalitis, (d) vascular lesions such as hemorrhage, embolism, thrombosis of veins and inflammatory lesions such as polioencephalitis. In the present case the anatomical defect is atrophy, due, as it appears to me, to a vascular defect or disease. It seems probable here that there was, during intra-uterine life, partial stoppage of the left middle cerebral artery, or there was a simple defect in the development of this artery and its branches, leading to the incomplete development of the brain which it nourished. The obliteration could not have been complete without leading to a more massive and localized lesion.

I do not think it possible for neurologists to tell from clinical studies exactly what is the organic anatomical condition in all cases of this kind. We can often be sure to detect a simple agenesis by the small size of the brain and the marked idiocy, but in this case the child's head was of fair size and the intelligence moderately good. We can tell the cause of other cases by the evidence of hydrocephalus and of still others by a distinct history of an acute onset of the disease subsequent to birth. By a process of exclusion we can therefore often reach a fairly accurate diagnosis. In the present case I was misled by the fact that the mother told us the attacks began with an acute illness several months after birth and it seemed possible that there had been an encephalitis or a hemorrhage with a possible formation of a cyst. This made the advisability of an operation a little greater, still even in cases where there seemed defective development it seems to me clinically to be shown that in some instances an operation upon the brain lessens the severity of the epileptic attacks and has a good influence in that way upon the development of the brain.